Misdiagnosis of persistent left superior vena cava with unroofed coronary sinus as a coronary sinus-type atrial septal defect

Alwaleed Al-Dairy[1], Reem Ahmad[1], and Rawan Hasan[1]

[1]Damascus University Faculty of Medicine

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Abstract

Unroofed coronary sinus syndrome may be associated with persistent left superior vena cava (PLSVC). Herein, we present a case of a 2-year-old patient who underwent an operation for repair of a coronary sinus-type ASD; however, PLSVC was detected intraoperatively. total repair was performed by creating an intra-atrial tunnel.

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Alwaleed Al-Dairy[1,[?]], MD, Reem Ahmad[2], Rawan Hasan [2].

[1] Assistant Professor in Cardiac Surgery at Faculty of Medicine, Damascus University, [2] Medical student Faculty of Medicine, Damascus University.

[*]**Correspondence to** : Alwaleed Al-Dairy: Assistant Professor in Cardiac Surgery at Faculty of Medicine, Damascus University, and Cardiac Surgeon at Damascus University Cardiac Surgery Hospital, Damascus, Syria.

Email: alwaleedaldairy.aa@gmail.com; Tel+963112133545, Fax +9632129437.

ORCID: Alwaleed Al-Dairy (0000-0002-2239-646X).

Abstract

Unroofed coronary sinus syndrome is a rare congenital heart defect representing less than 1% of all atrial septal defect types, and may be associated with persistent left superior vena cava which may be missed during preoperative diagnosis. Herein, we present a case of a 2-year-old patient who underwent an operation for repair of a coronary sinus-type ASD; however, PLSVC was detected intraoperatively. An antra-atrial tunnel was created to divert the flow of PLSVC into the right atrium along with the repair of the atrial septal defect.

Keywords : persistent left superior vena cava, unroofed coronary sinus, intra-atrial tunnel.

Key Clinical Message:

The association of UCSS with PLSVC must be kept in mind during operation. Creating an intra-atrial tunnel to divert the flow of PLSVC into RA, without obstructing the mitral valve or the pulmonary veins, is a safe surgical approach.

Introduction :

Persistent left superior vena cava (PLSVC) is the most common thoracic venous anomaly, with an incidence of 0.2% in normal neonates (1). However; it is more common among patients with congenital heart diseases

(CHD) (1.4%), and recognition of its presence by preoperative diagnosis is of a great importance during congenital heart surgery (1-4). Commonly, PLSVC drains into the right atrium (RA) via the coronary sinus (CS), and when isolated, it is usually asymptomatic, and may be detected incidentally (1). Unroofed coronary sinus syndrome (UCSS) is an uncommon CHD which leads to a left to right shunt at the atrial level, and comprises <1% of all atrial septal defect (ASD) types (2, 5). When UCSS is associated with PLSVC, it is difficult to detect the diagnosis by transthoracic echocardiography (TTE) (1). Herein, we present a case of a 2-year-old patient with preoperative diagnosis of coronary sinus-type ASD; however, PLSVC draining into the left atrium (LA) was detected during surgery.

Case presentation:

A 2-year-old girl was presented to our hospital when her parents noticed that she had suffered from moderate tachypnea recently. Physical examination revealed a 3/6 systolic murmur on the left sternal border and there was not any cyanosis. TTE showed an ASD of coronary sinus type of about 1.5 cm with dilated right heart cavities. The patient was scheduled for surgical closure of the ASD. Upon anaesthesia, oxygen saturation (SaO2) was 100%, and the operation was performed through median sternotomy. The pericardium was opened, and PLSVC was noticed. Complete cardiopulmonary bypass (CPB) was prepared without cannulating the PLSVC. The heart was arrested by antegrade cold blood cardioplegic solution. The right atrium (RA) was opened with an incision parallel to the right atrioventricular groove as usual. On inspection, we found that the PLSVC was draining into the roof of the LA, and a venous cannula was directed through the ASD towards its orifice to drain it (Figure 1). The pulmonary veins drainage and the mitral valve were inspected. A fresh autologous pericardial patch was used to construct a tunnel that drains the PLSVC into RA (Figure 2). The ASD was closed by another fresh autologous pericardial patch and thus the PLSVC will drain into RA (Figure 3). The remainder of the operation was completed uneventfully.

Discussion:

UCSS is a rare CHD representing less than 1% of all ASD types (1). Its clinical manifestation is atypical, and may be associated with varied CHDs (6). Preoperative diagnosis by TTE is very difficult when associated with PLSVC (2, 6). In our patient, the diagnosis of associated PLSVC was missed preoperatively, and was detected intraoperatively, and this had led to change in the surgical plan. Our surgical approach was to create an intra-atrial tunnel to divert the flow of PLSVC into RA with great attention to the pulmonary veins and the mitral valve, and repair the ASD with fresh autologous pericardial patch . Other operative options include: ligating PLSVC and repairing the ASD; constructing a baffle to guide PLSVC towards RA (6). We confirm the importance of precise preoperative diagnosis to avoid any complications from such an incidental intraoperative finding.

Conclusion:

UCSS is a rare CHD; however, its association with PLSVC must be kept in mind. Creating an intra-atrial tunnel to divert the flow of PLSVC into RA, without obstructing the mitral valve or the pulmonary veins, is a safe surgical approach.

Figure legends:

Figure-1: Intraoperative image showing the venous cannula placed through the ASD towards PLSVC orifice. 1: The opened RA, 2: LA, 3: the venous cannula placed into PLSVC orifice.

Figure-2: Intraoperative image showing the intra-atrial tunnel. 1: the pericardial patch used to construct the tunnel.

Figure-3: Intraoperative image showing the final anatomy after closing the ASD. 1: the pericardial patch used for ASD closure, 2: the venous cannula placed in the intra-atrial tunnel to divert PLSVC flow into RA.

Author Contribution

Alwaleed Al-Dairy : Planned and performed the work leading to the report. Wrote and reviewed successive versions and participated in their revisions.

Reem Ahmad : wrote and reviewed the successive versions and participated in their revisions

Rawan Hasan : Participated in writing the report and approved the final version

Author's Statement:

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The authors have no conflict of interest

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